Response to Letter Regarding Article, "Treatment of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia: An International Task Force Consensus Statement"

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We appreciated the interest of Barison and colleagues in our International Consensus Statement on the treatment of arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D).[1] Our document provided a comprehensive overview and recommendations for risk stratification and therapy of patients fulfilling the diagnostic criteria for ARVC/D. These criteria had been addressed by a previous International Task Force consensus document dedicated to diagnosis of ARVC/D.[2] According to the revised criteria proposed by the International Task Force in 2010, the diagnosis of ARVC/D is based on the presence of major and minor criteria encompassing electrocardiographic, arrhythmic, morphological, histopathologic, and genetic factors. Diagnosis of definite ARVC/D is fulfilled in the presence of 2 major criteria or 1 major plus 2 minor or 4 minor criteria from different groups; the ARVC/D diagnosis is possible or borderline in the presence of insufficient criteria.

In their Letter to the Editor, Barison and colleagues emphasized the valuable role of cardiac magnetic resonance (CMR) for diagnosis of ARVC/D, which relies on its ability to combine evaluation of ventricular size, function, and regional wall motion with characterization by late-gadolinium enhancement (LGE) of fibro-fatty myocardial scar, which is the hallmark lesion of ARVC/D. We totally agree that CMR is becoming the gold standard tool for detection of structural and functional ventricular abnormalities in ARVC/D. In the 2010 International Task Force consensus document, CMR was appropriately included among the diagnostic imaging techniques, and specific CMR reference values for normal (and abnormal) ventricular size, systolic function, and regional dyssynergy were provided. Tissue characterization by LGE was not included among these diagnostic criteria because of the potential risk of misdiagnosis of ARVC/D related to the difficulty of assessing LGE at the level of the thin right ventricular wall and possible confusion with normal epicardial fat tissue. More recently, the frequent involvement by the disease of the left ventricular wall in the form of epicardial-mediomural LGE has been recognized. As pointed out by the authors, LV LGE may enhance sensitivity for early/minor or predominant left variants of the ARVC/D; however, its diagnostic accuracy remains to be established.

The prognostic role of CMR and, in particular, of postcontrast sequences for tissue characterization in patients with a diagnosis of definitive ARVC/D remains elusive. In our consensus statement on the treatment of ARVC/D, we performed a systematic review of outcome studies on ARVC/D available in the literature to identify predictor variables that were associated with an increased risk of major arrhythmic events (ie, sudden cardiac death, appropriate defibrillator interventions, or defibrillator therapy on fast ventricular tachycardia/ventricular fibrillation), nonsudden cardiac death, or heart transplantation in at least 1 published multivariable analysis. Although the predicting value of CMR for worse arrhythmic outcome was shown in the general population of patients showing premature ventricular beats with a left bundle branch morphology[4] and in patients with suspected diagnosis of ARVC/D,[5] no specific features of CMR, either alone or in combination, have been reported yet as independent predictors of life-threatening arrhythmic events in patients with a definite diagnosis of ARVC/D. This lack of evidence underscores the importance of undertaking future studies on this field.

Disclosures

None.

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